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Case Report

Vulvar yolk sac tumor mixed with embryonal carcinoma in a peri-pubertal girl: A case report

Cherry Yin-Yi Chang [a,d,e], Kang-Hsi Wu [c], Tze-Yi Lin [b,e], Wei-Chun Chang [a,d], Wu-Chou Lin [a,d], Yao-Ching Hung [a,d], Lian-Shung Yeh [a,d,*]

[a] *Department of Obstetrics and Gynecology, China Medical University Hospital, Taichung 403, Taiwan*
[b] *Department of Pathology, China Medical University Hospital, Taichung 403, Taiwan*
[c] *Department of Pediatrics, China Medical University Hospital, Taichung 403, Taiwan*
[d] *College of Medicine, School of Medicine, China Medical University, Taichung 403, Taiwan*
[e] *College of Public Health, China Medical University, Taichung 403, Taiwan*

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Abstract

Objective: Vulvar cancer is rare in Eastern females, especially in pre- and peripubertal girls. The prognosis of vulvar cancer is poor and treatment is variable.

Case Report: A 14-year-old girl suffered from a left vulvar tenderness mass and underwent excision of the mass. The diagnosis of the pathology was vulvar yolk tumor with an embryonal carcinoma. After the vulvectomy, inguinal area lymph node dissection, chemotherapy (bleomycin, etoposide and cisplatin) treatment and radiotherapy, metastasis to lung was also noted after eight months. Resection of lung tumor was performed. She received chemotherapy with a combination of paclitaxel, ifosphamide and cisplatin (TIP) and received peripheral blood stem cell transplantation (PBSCT) twice and chemotherapy treatment of gemitabine and oxaliplatin (GEMOX). Up until now, the patient has been free of disease.

Conclusion: High-dose TIP and GEMOX chemotherapy plus PBSCT for bone marrow rescues could be considered to treat patients with metastatic malignant vulvar germ cell tumor after failed first-line chemotherapy and radiation.

Keywords: Embryonal carcinoma; Peripheral blood stem cell transplantation; Vulvar mass; Yolk sac tumor

Introduction

Vulvar yolk sac tumor is a rare but highly malignant tumor. To our knowledge, there were only 11 cases reported in literature until now [1–5]. But among these cases, there was only one case of vulvar yolk sac tumor combined with an embryonal carcinoma reported in 1978. There was a variety of treatment modality in these cases due to the rarity of vulvar yolk sac tumor. In this report, a girl with vulvar yolk sac tumor, an embryonal carcinoma and a lung metastasis with a stable condition after a combination treatment of surgery, chemotherapy, radiation and autologous peripheral blood stem cell transplantation (PBSCT) was presented.

Case report

A 14 year-old female student with progressive enlargement of left tender vulvar mass in one month was referred to University Hospital for gynecologic evaluation in September 2007. On physical examination, a 5 × 4 cm tender mass with well margin was located at left major labia and no palpable inguinal lymph node was found. The personal and family history of the patient was negative. This patient had normal development of breast, axillary and pubic hair. Excision of the mass was performed under intravenous general anesthesia in

* Corresponding author. 2Yuh-Der Road, Taichung 403, Taiwan.
E-mail address: lsyeh@mail.cmuh.org.tw (L.-S. Yeh).

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September 2007. Histological examination of the mass revealed a mixed germ cell composed yolk sac tumor (85%) and an embryonal carcinoma (15%) (Figs. 1–4). However, the tumor was present on the cut surface of the excision. After excision, a laboratory test showed her AFP level was 25.8 ng/mL, β-HCG, LDH and CA-125 were within normal limits and she had a normal 46XX karyotype. Positron emission tomography (PET) scan showed increased [18F]-2-fluoro-deoxy-D-glucose (FDG) uptake in the vulvar region. Magnetic resonance imaging (MRI) revealed several enlarged lymph nodes in the bilateral inguinal region. The patient received three courses of BEP chemotherapy (bleomycin 15 mg/m^2 iv weekly, etoposide 100 mg/m^2 and cisplatin 20 mg/m^2 Day 1 to 3 for both) given at 21-day intervals. AFP returned to normal limits in November 2007. After completion of three courses of chemotherapy, simple vulvectomy and bilateral inguinal lymph node dissection were performed. In the pathology report there was no residual tumor and no lymph node involvement. The patient was followed up monthly at OPD. Until July 2008, AFP was elevated to 12.36 ng/mL. MRI showed a 1.5 cm lymph node in left inguinal region and PET scan showed focal intensely increased FDG uptake in the left vulvar, left posteromost upper intrapelvic region and the inguinal region. After counseling with a radiation oncologist, radiation therapy with total dose 60 Gy/30 Fr were completed in August 2008. AFP returned to 0.98 ng/mL after radiation. However, one year after radiation APF elevated to 11.14 ng/mL. A chest X-ray showed a nodule over the left lower lobe (LLL) of the lung. MRI and computed tomography (CT) scan showed a 1.6 cm nodule in the lateral basal segment of the LLL of the lung. Tl-201 cancer work-up found an increased tracer uptake in the lung and perineal region. The patient received thoracoscopic wedge resection of the LLL of the lung and mediastinal lymph node dissection in September 2009. The pathology report revealed metastatic yolk sac tumor with an embryonal carcinoma and one lymph node was involved. In

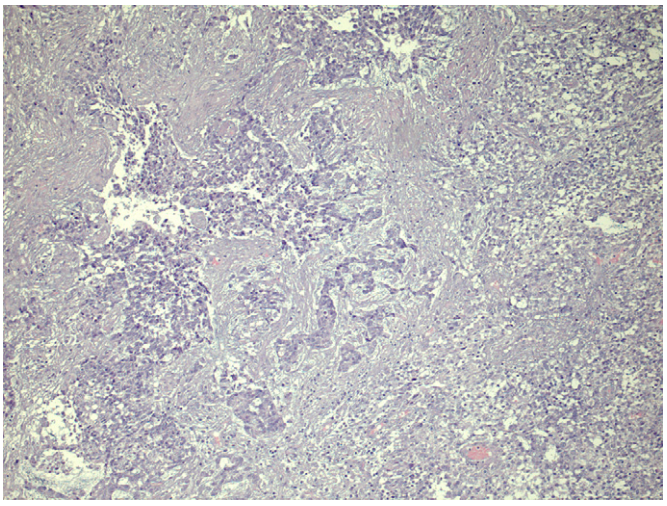

Fig. 2. Yolk sac tumor (right upper) revealing solid, microacinar, trabecular growth pattern and embryonal carcinoma (left lower) disclosing solid or glandular architecture with vesicular nuclei and prominent nucleoli, presence of foci of peritumoral intravascular microthrombi of both components, mild to moderate lymphoplasmacytic tumor-host reaction. (100×, H&E stain).

October 2009, the AFP level increased to 17.06 ng/mL. The patient was administered four courses of high-dose TIP chemotherapy (paclitaxel 250 mg/m^2 over 24 hours on Day 1, followed by ifosfamide 1500 mg/m^2 and cisplatin 25 mg/m^2 per day on Days 2 to 6) at 21-day intervals. Chemotherapy was completed in December 2009 and AFP subsequently decreased to 1.33 ng/mL. The patient received PBSCT in February and March 2010 and three courses of GEMOX chemotherapy (gemcitabine 1500 mg/m^2, intravenously on Days 1 and 8 and oxaliplatin 130 mg/m^2 on Day 8 every 21 days). After treatment, the patient had follow-up of a FDG PET/CT whole body scan which did not show any focally hypermetabolic lesions. Up until now, this patient has been well and the tumor marker is normal.

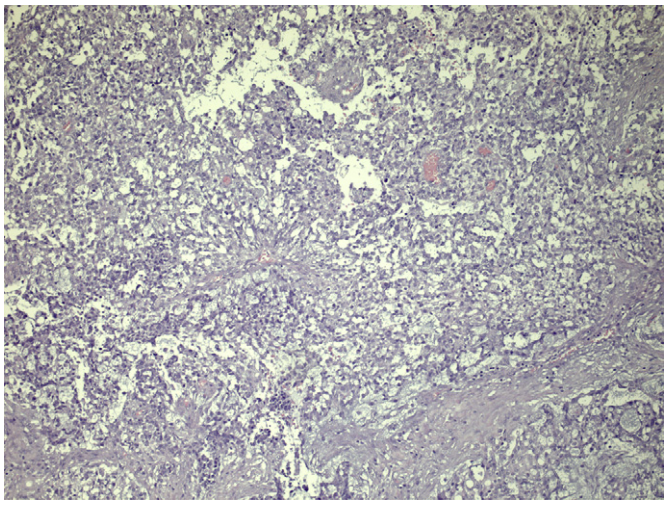

Fig. 1. Presence of a spider-weblike network formed by vacuolated cytoplasm of tumor cells, similar to a honeycomb (100×, H&E stain).

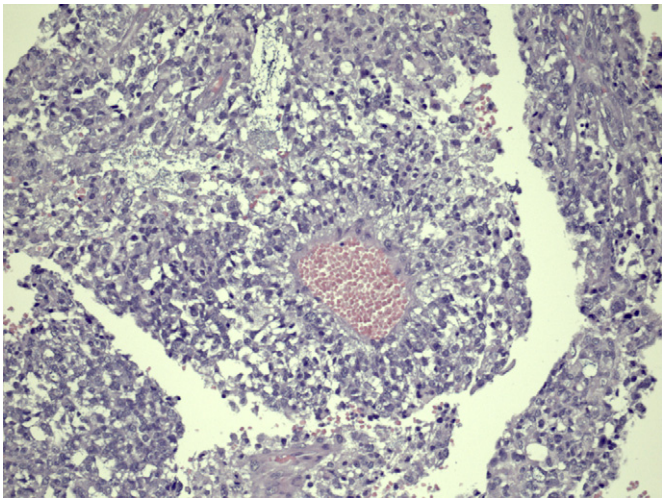

Fig. 3. Schiller-Duvall body characterized by the presence of a central vessel, surrounded by fibrous tissue and epithelial tumor cells, in a space lined by flat tumor cells. (200×, H&E stain).

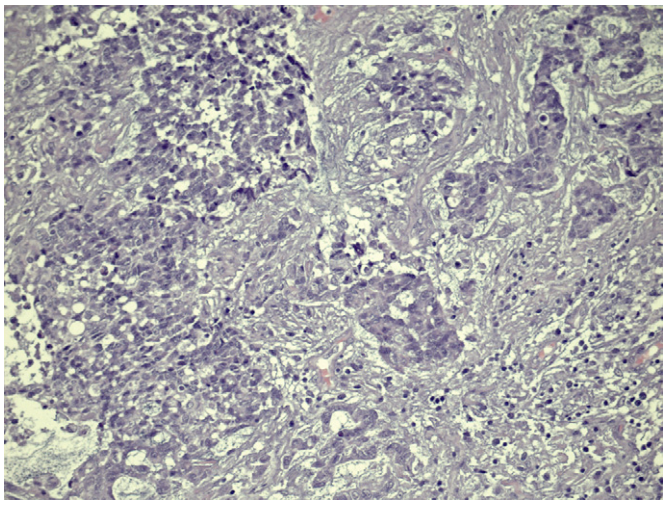

Fig. 4. Embryonal carcimoa showed glandular pattern with sheets, glands and nests of malignant large ovoid or polygonal cells with pleomorphic nuclei. (200×, H&E stain).

Discussion

Vulvar tumor is rarely seen in girls but it is important to make a differential diagnosis of benign tumor or malignancy. Most benign vulvar masses have well defined margins without tenderness and potentially malignant masses are not well defined and the patient could have symptoms of tenderness [6]. In this case, the patient presented signs of an infection and had antibiotic treatment at clinics which failed so she was referred to the University Hospital for management.

Malignant vulvar germ cell tumor is rare and the majority are pure yolk sac tumors. The commonly used treatment for malignant vulvar yolk sac tumor is conservative surgery (local or wide excision combined with/without inguinal lymph node sampling or dissection) followed by adjuvant chemotherapy. The chemotherapy traditionally used in treating germ cell tumor was BEP, PVB (cispltain, vincristine and bleomycin) or VAC (vincristine, dactinomycin and cyclophosphamide). However, the regimen is commonly used in the treatment of ovarian cancer and some patients had resistance to a cisplatin-based regimen in treating vulvar cancer [7]. In this case, the patient accepted BEP treatment first but the tumor marker was elevated and the recurrence noted at the inguinal area lymph node within one year after chemotherapy. In previous reports, most recurrent or metastatic sites were the inguinal lymph node or pleura/lung and recurrence mainly occurred within two years. Adjuvant radiation therapy was the choice to treat recurrence but three cases died within two years, so treatment for this patient with recurrence was a critical issue. Kondagunta et al study showed 65% of patients with relapsed testicular germ cell tumor had two-year progression-free survival after the second-line high-dose TIP chemotherapy [8]. Mead et al study also revealed the good response with high-dose TIP treatment for patients with metastatic malignant germ cell tumor post BEP chemotherapy [9]. However, salvage high-dose chemotherapy led to 64–70% patients with grade 3 to 4 leucopenia and 7% had grade 4 to 5 renal toxicity. Autologous bone marrow cells transplantation has been reported as benefiting patients by rescuing the hematopoietic system after high-dose chemotherapy. In this case, the patient accepted PBSCT twice for rescue. The patient then received GEMOX chemotherapy treatment which was used in patients with non-small lung cancer and both refractory or sensitive ovarian cancer and had moderate and tolerable side effects and toxicity [10,11]. During the three courses of GEMOX chemotherapy, this patient did not suffer pancytopenia or severe nausea and vomiting.

In conclusion, a vulvar mass of a young girl is rare and it is important to make correct diagnosis. High-dose TIP and GEMOX chemotherapy plus PBSCT for rescues could be considered to treat patients with metastatic malignant or recurrent vulvar germ cell tumor after failed first-line chemotherapy and radiation.

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